

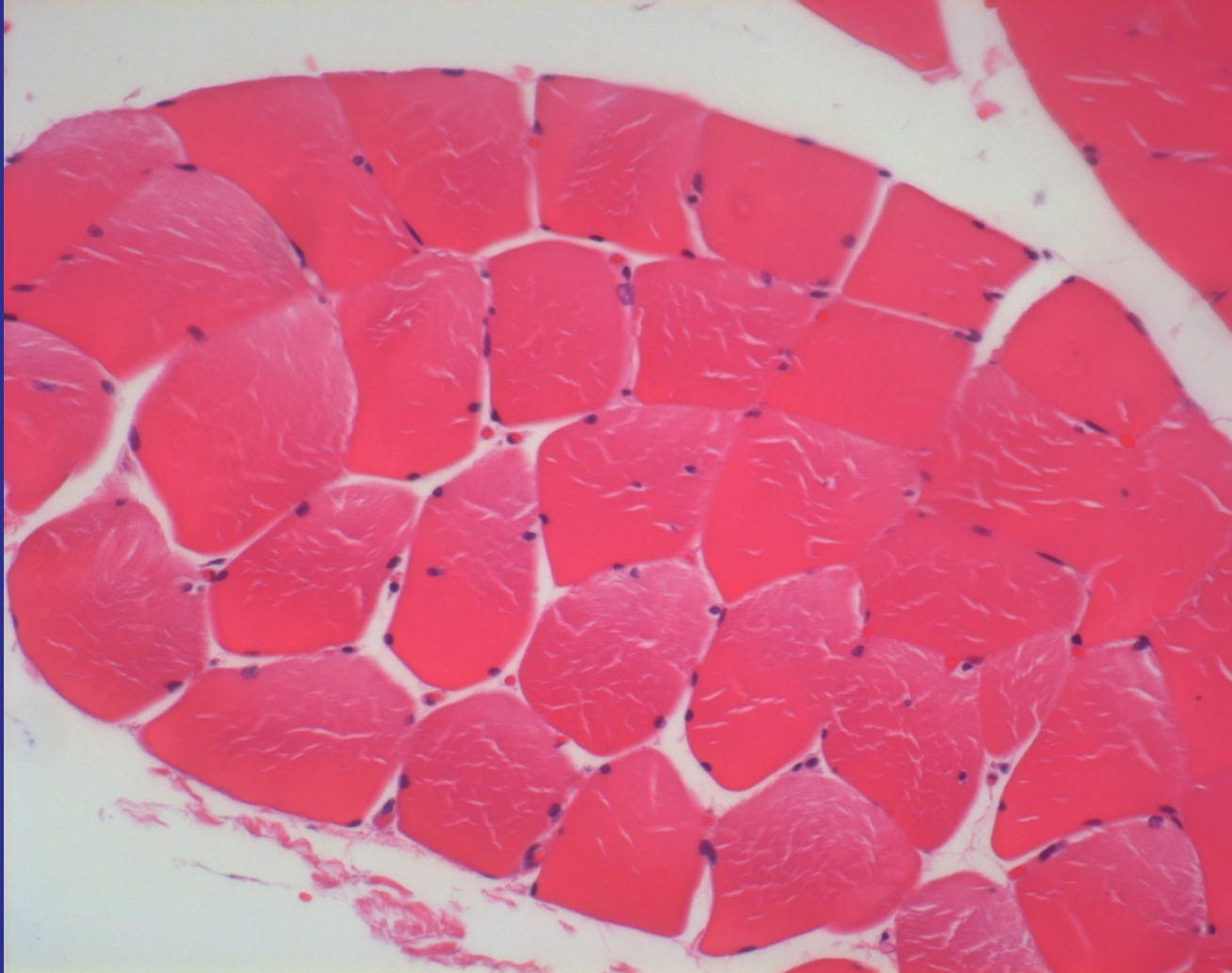
IBM: Is it an inflammatory disease or am I just getting old?

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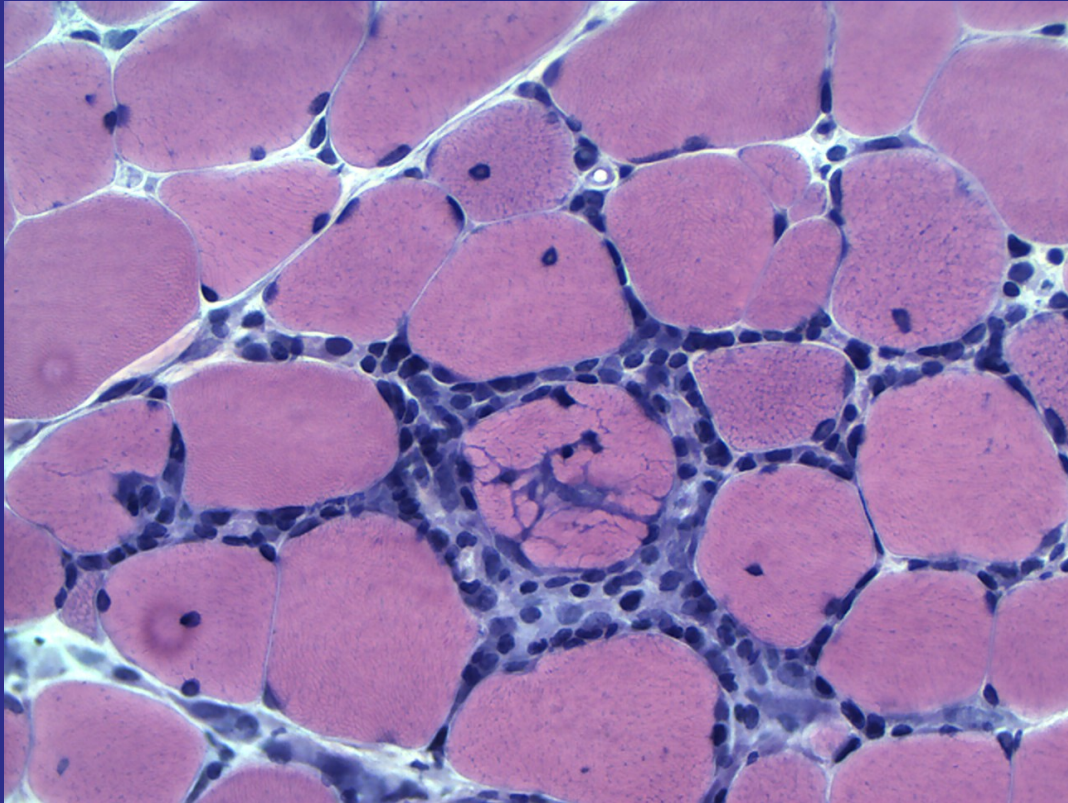
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Normal Muscle

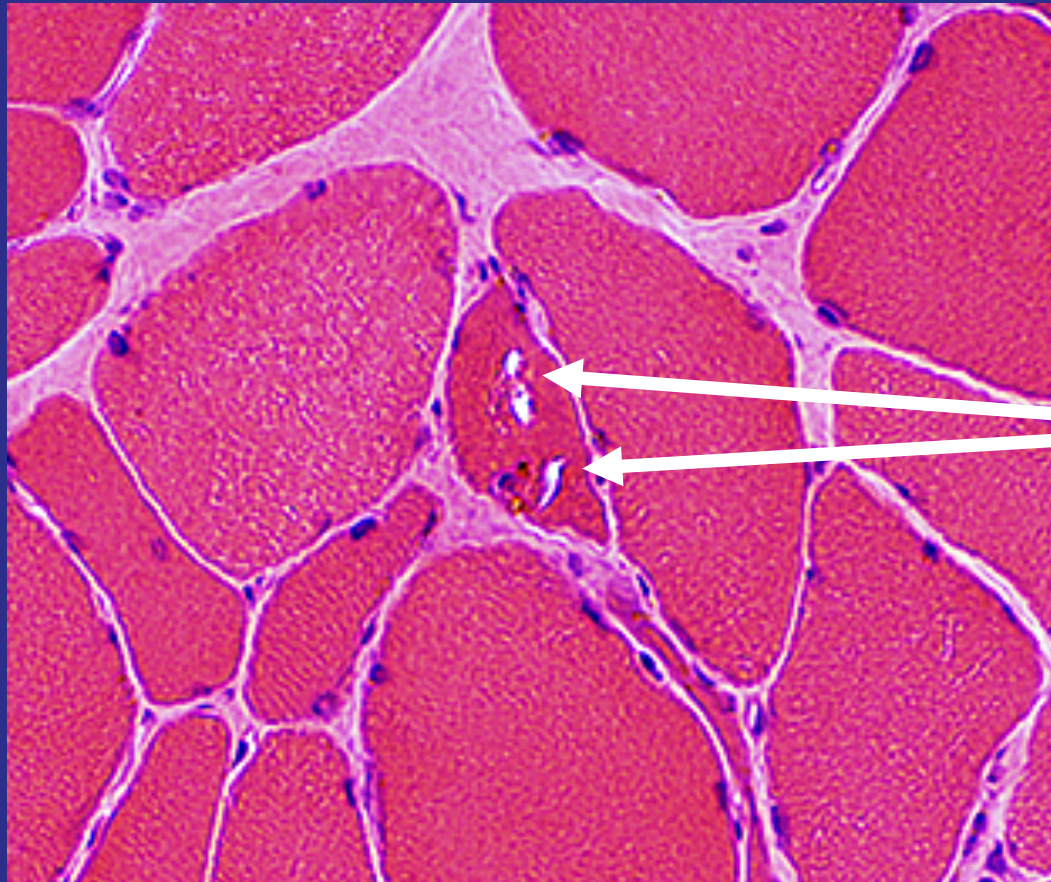


Autoimmune myositis (i.e., polymyositis) muscle



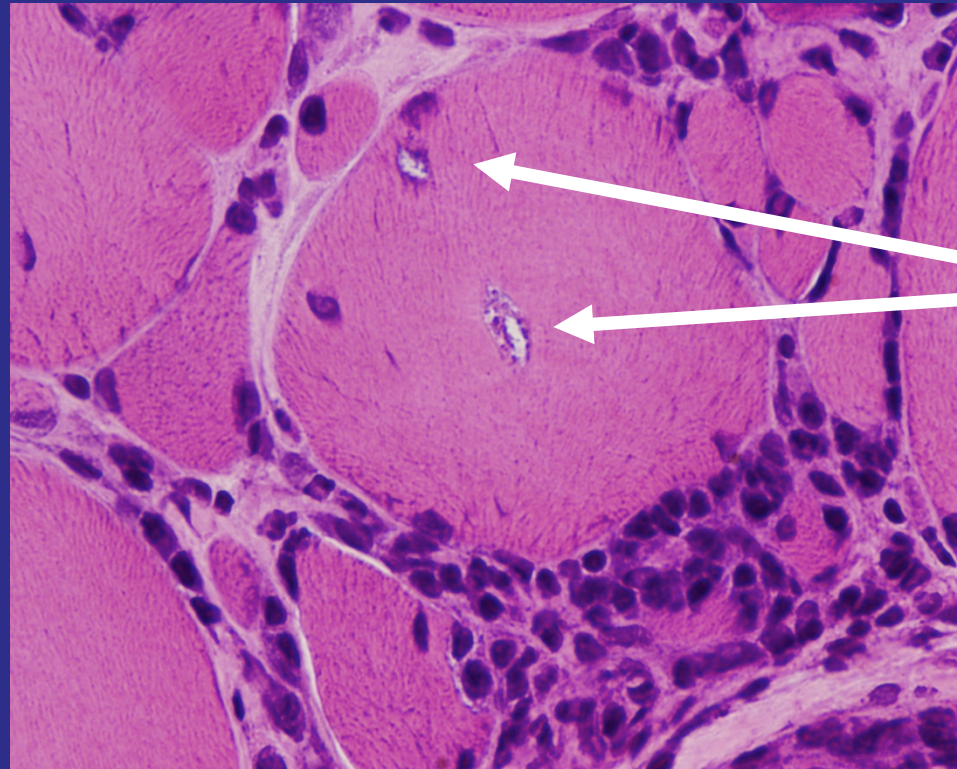
Muscle cells surrounded and invaded by inflammatory cells (CD8+ T cells)

Genetic muscle disease (oculopharyngeal muscular dystrophy) muscle



Rimmed vacuoles

Inclusion Body Myositis



Rimmed vacuoles

Figure courtesy of Dr. Amato

Inflammation and rimmed vacuoles

Inclusion body myositis

- Degeneration?
 - Vacuoles contain abnormally folded proteins
 - Some patients who look like they have sIBM have genetic mutations (e.g., VCP mutations)
 - No response to immunosuppression
- Autoimmune?
 - Very aggressive inflammatory cells (CD57+ T cells) identified in blood and muscle
 - Autoantibodies identified

Approaches to treat IBM

- Prevent/improve abnormal protein folding and accumulation (as seen in rimmed vacuoles)
 - E.g., arimoclomol
- Immune suppression
 - Traditional drugs
 - Drugs designed to target the autoaggressive T cells in IBM
- Make existing muscle stronger
 - E.g., Bimagrumab
 - Exercise
- Combination therapy